Blood disorders:

1. The single best test to confirm iron deficiency is?
   - A. Plasma iron
   - B. Plasma ferritin
   - C. Iron binding capacity
   - D. Peripheral film
   - E. Transferrin saturation

Ans: Plasma ferritin

Explanation: Plasma ferritin is a measure of iron stores in tissues and is the best single test to confirm iron deficiency. It is a very specific test; subnormal level is due to iron deficiency or very rarely hypothyroidism and vitamin C deficiency. Ferritin levels can be raised in liver disease and in the acute phase response. Plasma iron and total iron binding capacity are measure of iron availability hence they are affected by many factors besides iron stores. Plasma iron has a day-to-day variation and becomes very low during an acute phase response and is raised in liver disease and hemolysis. A transferrin saturation of less than 16% is consistent with iron deficiency but is less specific than a ferritin measurement. Iron deficiency causes microcytosis and hypochromia but it is also not specific for IDA.

2. A patient has history of gut surgery which included partial gastrectomy 3 months back now presented with fatigue and shortness of breath on exertion. CBC done which showed hemoglobin of 10. What is the most probable cause of anemia in this patient?
   - A. Iron deficiency
   - B. Vitamin B12 deficiency
   - C. Folate deficiency
   - D. Chronic blood loss
   - E. Anemia of chronic disease

Ans: Iron deficiency

Explanation: Iron deficiency anemia is the most common cause of anemia after partial gastrectomy. Iron deficiency after gastrectomy or bypass results from malabsorption of iron because of a surgically altered gastrointestinal tract. Inadequate oral intake or occult blood loss may also contribute to this condition. Iron is absorbed in the duodenum and proximal jejunum and its absorption is enhanced by gastric acid secretion. A lack of acidity results in impaired conversion of ingested ferric iron to absorbable ferrous iron. Duodenal bypass also contributes to iron deficiency in patients with gastrectomy or bypass surgery. Anemia due to Vitamin B12 level occurs after 2 to 3 years post operation. It commonly occurs with total gastrectomy.

3. A 23 year old male presented with feeling low energy in body with tiredness all the times. CBC done which showed HB of 9.1 with MCV of 65. WBC are slightly increased with high eosinophil count. What is the probable cause of anemia in this patient?
   - A. Blood loss
   - B. Celiac disease
   - C. Hookworm infestation ✓ 65
   - D. Vitamin B12 deficiency
   - E. Thalassemia

Questio.

Ans: Hookworm infestation

Explanation: Low HB with microcytosis suggests iron deficiency most probably. High eosinophil count points towards parasitic infestation which is the most common cause of iron deficiency in developing countries.

4. A 27 year old female is presented with complaints of intermittent dysphagia. Examination showed marked pallor, angular cheilitis with glossitis. What initial treatment should be given to her?
   - A. Vitamin supplements
   - B. Iron supplements ✓
   - C. Esophageal dilation
   - D. Esomeprazole
   - E. Folic acid supplements

Ans: Iron supplements

Explanation: This is a case of Plummer Vinson syndrome which consists of classical triad of dysphagia, iron deficiency and esophageal webs. It is also associated with cheilitis and atrophic glossitis. Initial treatment is with iron supplements and if symptoms of dysphagia persists after iron supplements then mechanical dilation is done. It is associated with increased risk of [illegible] esophagus.

5. A 25 year old female is diagnosed as having iron deficiency anemia with a hemoglobin of 10 and ferritin of 20. She is started on oral iron supplements. What should be the minimum duration of oral therapy?

15-300
100-200%

- A. 1 month
- B. 2-3 months
- C. 3-6 months ✓
- [✓ D. 3 months after normalization of ferritin]
- E. 1 year

1.0g/dL
2
3
>160
>20

Ans: 3 months after normalization of ferritin

Explanation: oral iron supplements should be given at least 3 months after hematocrit and ferritin become normal to restore the iron stores in the body.

6. Which one of the following parameters starts getting better earliest after starting iron supplements for iron deficiency anemia?

- A. Increase in hemoglobin 2 3-
- B. Increase in ferritin
- [✓ C. Increase reticulocyte count]
- D. Feeling of wellbeing
- E. Normalization of cell size

7-12
18
4
1.0g/L Hb.

1.0g/L /week
1 months
7.08
8-10.12
5-7
MDS
1g/L /week
2 6 2-3 2 6

Ans: Increase reticulocyte count

3

Explanation: Hemoglobin starts improving after 2 weeks, reticulocyte count increase after 5-7 days of treatment, ferritin also increased after 2 weeks. Patient sense of wellbeing starts improving after 1 month. The best predictor is improvement in fatigue. An increase of > 1 g in hemoglobin after 2 weeks is the best follow up test to confirm the response of oral iron. If less than 1 g with no ongoing loss of iron, shift to IV iron therapy.

2-1.0g/L

7. A 50 year old male known case of heart failure with EF of 30% on standard medical therapy complains of fatigue and worsening SOB on exertion. Labs were done which showed an Hb of 9.8 with ferritin of 21 and transferrin saturation of 15%. What should be the next best management?

- A. Oral iron supplements
- B. IV iron therapy
- C. Erythropoietin

100
20
>100
>20

D. Whole Blood transfusion

E. PRBC transfusion

Ans: IV iron therapy

Explanation: History and lab results confirm iron deficiency anemia in a patient with heart failure. Iron deficiency anemia is widely present in patients with heart failure with an estimated prevalence of over 50% in ambulatory patients. It is an independent predictor of worse functional capacity and survival. Definition of iron deficiency in heart failure is as follow: ferritin <100 µg/L or ferritin of 100-299 µg/L with a transferrin saturation <20%. Intravenous (IV) iron is the preferred route for treatment in heart failure patients. IV iron administration in HF is associated with improvement in patient-reported outcomes and functional capacity. The efficacy of oral iron in patients with heart failure with reduced ejection fraction is poor. Furthermore, oral iron preparations are associated with a high incidence of adverse effects, are poorly absorbed due to gut wall edema, and can take up to 6 months to replenish iron stores. Erythropoietin therapy is not recommended in heart failure. PRBC transfusion is only used in emergency cases.

8. Which electrolyte disturbance is most common after treatment with IV iron therapy?

A. Hyponatremia

B. Hypokalemia IV

C. Hypophosphatemia ✓

D. Hyperkalemia

E. Hypocalcemia ✓

Ans: Hypophosphatemia

Explanation: Iron infusion therapy is used to treat severe iron deficiency which is not responding to the first-line oral iron therapy. It can cause severe renal wasting of phosphate resulting in severe hypophosphatemia in some patients. Pre-existing vitamin D deficiency, low calcium levels, low phosphate levels or raised parathyroid hormone levels are the risk factors, and these should be evaluated and corrected before administering intravenous iron. The common symptoms are muscle cramps and weakness with fatigue. In severe deficiency, it can also presents with respiratory muscle weakness. Treatment is with vitamin D and phosphate supplements.

6-12

9. A known case of CKD stage 3b is diagnosed as having iron deficiency anemia with hemoglobin of [illegible] started on ferrous sulfate 3 months back. Patient has good compliance to treatment. Hb done

today is 10.2 with a ferritin of 40 and transferrin saturation of 16%. What should be the next treatment?

A. Increase dose of oral iron

B. Start IV iron ✓

C. Erythropoietin

D. PRBC transfusion

E. Continue same dose of oral iron

7.0-8
10.0 g/L
50,00

Ans: Start IV iron

Explanation: Iron deficiency anemia is a common complication of chronic kidney disease (CKD). CKD patients suffer from both absolute and functional iron deficiency. Absolute iron deficiency is defined by severely reduced or absent iron stores, while functional iron deficiency is defined by adequate iron stores but insufficient iron availability for incorporation into erythroid precursors. This is due to increased levels of hepcidin. Anemia in CKD is associated with an increased risk of morbidity and mortality. The association between anemia and mortality may be related to the severity of anemia. All CKD patients should be screened for anemia during the initial evaluation for CKD. Criteria used to define iron deficiency are different among CKD compared to normal renal function. Among CKD patients, absolute iron deficiency is defined when the transferrin saturation is ≤20% and the serum ferritin concentration is ≤100 ng/mL among predialysis and peritoneal dialysis patients or ≤200 ng/mL among hemodialysis patients. Functional iron deficiency, also known as iron-restricted erythropoiesis, is characterized by transferrin saturation ≤20% and elevated ferritin levels. Iron supplementation is recommended for all CKD patients with anemia. There is general agreement according to guidelines that intravenous iron supplementation is the preferred method for CKD patients on dialysis (CKD stage 5) and either IV or oral iron is recommended for patients with CKD stages 3–5 not on HD. Guidelines suggest using oral iron for individuals who are not on erythropoietin and offering IV iron to those who do not tolerate oral therapy or do not reach targets within 3 months. For those receiving erythropoietin, the IV route is preferred. As this patient is still iron deficient after three months of oral iron, next best step should be IV iron. If after IV iron hemoglobin target is not achieved, erythropoietin should be given.

2.0g/L

[illegible]

1.0g/L

2.0g

10. A 51 year old male is investigated for exertional dyspnea diagnosed as having iron deficiency anemia. What should be the next best management step for him?

② 1g/dL

A. Upper GI endoscopy

B. Colonoscopy

② months ①

C. Fecal occult blood

D. CT abdomen

E. Urine routine examination

Screening test ⇒ tumours

Ans: Colonoscopy

Explanation: Iron deficiency anemia in male of all ages and post-menopausal women is due to gastrointestinal bleeding until proven otherwise and it should be investigated with colonoscopy followed by upper GI endoscopy. Fecal occult blood is not sensitive and specific and is not recommended.

11. What is the most common abnormality present on peripheral film in patient with anemia of chronic disease?

    A. Microcytic, hypochromic

    B. Microcytic, normochromic

    C. Normocytic, normochromic

    D. Normocytic, hypochromic

    E. Microcytic, hyperchromic

m

1 5 6

3 5. 8 10-12

1.0 g/12 Hb

Ans: The typical anemia of chronic disease is normocytic and normochromic. It can becomes microcytic hypochromic as the diseases progresses.

12. A 35 year old female is investigated for the cause of anemia. Labs showed Hb 10.2, WBC 9000, platelets 345,000. MCV 71, MCH 29, low reticulocyte count, ferritin level of 350 with low serum iron. Further tests showed high hepcidin level. What is the diagnosis?

    A. Iron deficiency

    B. Anemia of chronic disease ✓

    C. Folate deficiency

    D. Sideroblastic anemia

    E. Hemolytic anemia

ferritin

>100

>20

Ans: Anemia of chronic disease.

Explanation: Anemia of chronic disease is characterized by normochromic normocytic blood picture with low serum iron, decreased transferrin saturation, decreased bone marrow Sidroblasts and increased reticuloendothelial iron with normal or increased ferritin. Hepcidin level is typically elevated. The mechanism that produce the anemia include impaired production of erythropoietin, blunted marrow erythroid response to erythropoietin, iron restricted erythropoiesis and diminished pool of erythropoietin responsive cells. It is associated with chronic diseases and infections or neoplasia. Low serum iron with high ferritin and high hepcidin level are typical of anemia of chronic disease. The gold standard test is bone marrow aspiration with iron staining.

13. A bone marrow aspiration of the patient is done for the evaluation of unexplained anemia which showed 20% of nucleated erythroid cells containing ring sideroblasts. Which treatment should be given to the patient?

    A. Iron supplements
    B. Vitamin B12
    C. Folic acid
    D. Pyridoxine
    E. Pyridoxine + folic acid

1.0g

pyri

Ans: Pyridoxine + folic acid

Explanation: This is a case of Sideroblastic anemia. Pyridoxine and folic acid should be given to all patients with this diagnosis on bone marrow examination. The Sideroblastic anemias are a group of inherited and acquired bone marrow disorders characterized by pathological iron accumulation in the mitochondria of erythroid precursors. Abnormal, iron-laden mitochondria appear to encircle erythroblast nuclei, giving rise to the defining morphological feature of the ring sideroblasts.

14. Blood film of a patient shows macrocytosis with poikilocytosis and neutrophils with 6 lobes. What is the probable diagnosis?

    A. Alcoholism
    B. Hypothyroidism
    C. Folate deficiency
    D. Liver disease
    E. Iron deficiency

1.0 g/dl

2 week

1 month

pellagra

Ans: Folate deficiency

Explanation: All are the causes of macrocytosis except iron deficiency. Hyper segmented neutrophils are present in Folate or B12 deficiency. Liver disease causes target cells with stomatocytes, alcohol and hypothyroidism only cause macrocytosis.

15. Which one of the following is the earliest neurological manifestation of cobalamin deficiency?

    A. Motor weakness

    B. Paresthesia in extremities ✓

    C. Ataxia

    D. Dementia

    E. Sphincter disturbance

Ans: Paresthesia in extremities

Explanation: Cobalamin deficiency is associated with neurological disease in up to 40% of cases. The main pathological finding is focal demyelination affecting the spinal cord, peripheral nerves, optic nerves and cerebrum. The most common manifestation is sensory with peripheral paresthesia followed by ataxia of the gait.

16. A pregnant female has work up for low hemoglobin. Labs showed Hb 10.2, MCV of 61 with normal platelets and RDW value. Total RBC count is also high. Patient is otherwise asymptomatic. What is the most probable cause of anemia?

    A. Iron deficiency

    B. Beta thalassemia trait ✓ 10

    C. Beta thalassemia major

    D. Sideroblastic anemia

    E. Folate deficiency

Ans: Beta thalassemia trait

Explanation: The two main differential in this patient who is asymptomatic and having microcytic anemia are iron deficiency and beta thalassemia trait. The clue that this is beta thalassemia trait rather than iron deficiency is that the MCV is surprisingly low for the level of hemoglobin. Additionally, total RBC count is also high with normal RDW value which also points towards thalassemia trait rather than iron deficiency. In iron deficiency, RBC count is low and RDW is high. The next test should be hemoglobin electrophoresis which will show increase HbA2 value.

30. [illegible] 8

17. A 23 year old male has malaria which was treated with primaquine 4 days back. Now patient presented with jaundice and fatigue. Labs showed Hb of 8.1 with normochromic normocytic picture, bilirubin 4.1, ALT 24 and alkaline phosphatase 102. G6PD levels are normal. Peripheral film also showed bite cells. What is the probable diagnosis?

A. Plasmodium falciparum malaria

B. Mycoplasma infection

C. G6PD deficiency ✓ Heinz

D. Hereditary spherocytosis

E. Dublin Johnson syndrome

Ans: G6PD deficiency

Explanation: This patient is having acute hemolysis. Bite cells and blister cells with Heinz bodies are diagnostic of G6PD deficiency. Patient has taken primaquine which also causes acute hemolysis in G6PD deficient patients. During acute hemolysis, G6PD levels are falsely normal. Malaria will cause hemolysis with altered LFTs.

18. Aplastic sickle cell crisis is caused by?

A. Mycoplasma

B. HIV

C. Parvovirus B19 ✓

D. Cytomegalovirus

E. Epstein Barr virus

Ans: Parvovirus B19 causes aplastic crisis in sickle cell patients.

19. A diagnosed case of sickle cell disease patient presented with history of sudden onset of left sided weakness for 2 hours. Patient has history of diarrhea 2 days back. CT brain is normal. What is the best treatment for this patient?

A. Thrombolysis

B. Aspirin

C. Plasmapheresis

D. Clopidogrel

E. Exchange transfusion

TTP.

Vaso

Aplas

SLE.

Hemo

R

Musk

Pregi plasma

Ans: Exchange transfusion

Explanation: This is a case of cerebral vaso-occlusive episode as a result of sickle cell disease. The treatment of choice in this situation is urgent and aggressive exchange transfusion followed by transfusions to maintain HbS less than 30%. Thrombolysis, aspirin and clopidogrel has no role in this patient.

20. A patient is diagnosed as having megaloblastic anemia. Result of B12 and Folate are awaited. You want to start treatment. Best step in treatment would be:
    - A. Folic acid before B12
    - B. B12 before folic acid
    - C. Folic acid only
    - D. B12 only
    - E. B12 and Folate simultaneously

Ans: B12 and Folate simultaneously

Explanation: There is a risk that if folic acid is given to people who have undiagnosed deficiency of vitamin B12 it may lead to neurological damage. Vitamin B12 deficiency produces both an anemia identical to that of folate deficiency but also causes irreversible damage to the central and peripheral nervous systems. Folic acid will correct the anemia of vitamin B12 deficiency and so delay diagnosis but will not prevent progression to neurological damage. For this reason, folic acid supplements may involve simultaneous administration of vitamin B12.

21. Which electrolyte should be monitored during treatment with Folate and cobalamin therapy?
    - A. Sodium
    - [✓ B. Potassium]
    - C. Calcium
    - D. Phosphate
    - E. Chloride

Am

R

Ans: Potassium

Explanation: Serum potassium levels can fall during therapy for severe cobalamin or folate deficiency as new cells are formed rapidly for which potassium is taken inside the cell. It can lead to sudden death. Therefore, potassium should be monitored and supplements may be indicated.

22. Morphological features of megaloblastic anemia can include all of the following except:
    - A. Oval macrocytes
    - B. Hyper segmented neutrophils

C. Hypo cellular marrow

D. Pancytopenia

E. Giant megakaryocytes

3 month 1 02 6

Ans: Hypo cellular marrow

Explanation: Bone marrow aspiration is usually not needed to make the diagnosis of megaloblastic anemia. However, it can help rule out myelodysplasia and check iron stores. The bone marrow is hyper cellular with erythroid hyperplasia. Erythroid precursors have megaloblastic features being larger than normoblastic cells. In addition nuclear maturation is immature relative to cytoplasmic maturation. Megaloblastic changes are most prominent in more mature RBC precursors. Giant bands neutrophil precursors and hyper segmented neutrophils are present. Megakaryocytes are large and hyper lobulated. These changes are reversed within 12 hours of treatment with cobalamin or folate and morphology will be normal after 2-3 days. Therefore, bone marrow aspiration if necessary should be performed before therapy.

5-7 02 3 months 1.0g/dl

23. A 25 year old female presented with weakness with anorexia and sore tongue for 3 months. Examination showed marked pallor with yellow sclera and smooth shiny tongue with multiple white color patches on skin. Which test will confirm your suspected diagnosis?

A. Peripheral blood smear

B. Serum Bilirubin

C. Intrinsic factor antibodies

D. Anti-parietal cell antibodies

E. Serum Folate level

cef Amp

Ans: Intrinsic factor antibodies

Explanation: On the basis of history and examination, in a patient with weakness and sore tongue with anorexia and vitiligo with jaundice, the most probable diagnosis is Pernicious anemia. All of the above tests are used in the evaluation of megaloblastic anemia but Pernicious anemia is confirmed with anti-Intrinsic factor antibodies. These antibodies are present in up to 50% of patients. Parietal cell antibodies are present in 90% of patients with Pernicious anemia but these are also present in 25% of normal population, so these are not specific and not used for confirmation.

24. Which one of the following is the most specific for intravascular hemolysis?

A. Increase Bilirubin

B. Increase urinary urobilinogen

C. Haemosiderinuria ✓

D. Increase LDH

E. Increase reticulocyte count

LDH
[illegible]
[illegible] ↑
Hepto

Ans: Haemosiderinuria

Explanation: Red cell destruction causes a rise in unconjugated bilirubin in the blood and mild jaundice due to hemolysis. Increased reabsorption of urobilinogen from the gut results in an increase in urinary urobilinogen. Red cell destruction releases LDH into the serum. The bone marrow compensates with reticulocytosis and nucleated RBC precursors appear in the blood. These features are present in all causes of hemolysis. In intravascular hemolysis, free hemoglobin is released into the plasma which bind haptoglobin, resulting in a fall in its level during active hemolysis. Once haptoglobin is saturated free hemoglobin is oxidized to form meth- hemoglobin. Meth-hemoglobin is degraded and any heme is bound to hemopexin. When all the protective mechanisms are saturated, free hemoglobin appears in the urine as hemoglobinuria. Renal tubular cells absorb the hemoglobin and store it as hemosiderin. When these tubular cells sloughed into urine, they give rise to Haemosiderinuria, which is the most specific finding of intravascular hemolysis.

25. A 35 year old lady admitted with acute calculous cholecystitis has blood tests which showed Hb of 10.1 with high MCHC. Examination showed splenomegaly. Retic counts were 7%. Direct and indirect coombs test are negative. What is the most probable cause of anemia in this patient?

A. Autoimmune hemolytic anemia

B. Malaria

C. Hereditary spherocytosis ✓

D. Thalassemia

E. Sickle cell disease

Ans: Hereditary spherocytosis

Explanation: Anemia with gall bladder stones with negative coombs test points towards chronic hemolytic anemia like hereditary spherocytosis, thalassemia and sickle cell disease. High MCHC is present in HS. Also sickle cell disease and thalassemia presents in young age. Anemia of HS is mostly detected incidentally. In HS, red cell membrane structure is disrupted, which leads to loss of elasticity of the RBCs. When these cells pass through spleen, they lose their membrane relative to their cell volume. This results in an increase in MCHC and spherocytosis. Pigment gallstones are present in more than 50% of patients and patient can present with cholecystitis.

26. A 15 year boy from Afghanistan presented with hepatosplenomegaly. Patient has history of multiple blood transfusions since childhood. Blood film is normocytic normochromic. What is the probable diagnosis?
   - A. Thalassemia major ✓
   - B. Thalassemia trait
   - C. Sickle cell disease
   - D. Hereditary spherocytosis
   - E. Anemia of chronic disease

Ans: Thalassemia major

Explanation: When two defective genes for the β-globin molecules are inherited, the patient is affected with thalassemia major, also known as Cooley's anemia. Thalassemia major is generally detected during the 1st year of life because severe microcytic hypochromic anemia develops as fetal hemoglobin synthesis ceases after 3-4 months of age. The consequently produced exceptionally fragile red blood cells can survive for only few days in the peripheral circulation, which leads to critically increased rate of hematopoiesis (up to 30 times the normal), in an attempt to maintain adequate oxygenation. This results in massive bone marrow hyperplasia, in association with hepatosplenomegaly and lymphadenopathy due to extra medullary hematopoiesis. Blood transfusion is the customary treatment for thalassemia major, administered every 2-3 weeks so as to simulate the normal hematologic state. As transfusion is done every 2 to 3 weeks, blood film is mostly normocytic normochromic due to transfused blood. Blood transfusion is not needed in thalassemia trait. In sickle cell, there is splenic atrophy. Hereditary spherocytosis generally not cause severe anemia which needs regular transfusion. Anemia of chronic disease is also mild anemia.

LDH
tuuan

27. A 49 year old female is presented with burning sensation in both feet associated with ataxia for 2 weeks. Examination showed a pale looking female with smooth tongue with loss of vibration sense. Labs showed macrocytosis and hyper segmented neutrophils. What should be the treatment duration for this patient?
   - A. 6 months
   - B. 3 months
   - C. 9 months

D. 12 months

E. Lifelong

Ans: Lifelong

Explanation: This is a case of neuropathy associated with vitamin B12 deficiency. The classical triad of pallor, sore tongue and paresthesia typically occurs due to Vitamin B12 deficiency. Onset is often with a sensation of cold, numbness, or tightness in the tips of the toes and then in the fingertips, rarely with lancinating pains. Paresthesia's are ascending and occasionally involve the trunk, leading to a sensation of constriction in the abdomen and chest. Untreated patients may develop limb weakness and ataxia. Loss of vibration sense occurs earliest. It can also cause up going plantar with absent ankle reflex. Early in the course, poor joint position and vibration sense predominate. Typically, the legs are affected before the arms. Rarely are all limbs affected simultaneously. A Romberg sign is commonly found. The gait may be wide based. On presentation, 50% of patients have absent ankle reflexes with relative hyperreflexia at the knees. Plantars are initially flexor and later extensor. Standard treatment in patients with vitamin B-12 deficiency consists of parenteral or oral cobalamin. The recommended regimen is cobalamin 1000 mcg IM/SC daily for 5 days followed by 1000 mcg/wk. for 5 weeks, then 100-1000 mcg/mo. for life. Because 1% of cobalamin is absorbed by passive diffusion, administration of large oral doses is an alternative; 1000 mcg daily yields a daily absorption of 10 mcg, which exceeds the 2-mcg recommended daily allowance requirement.

28. A patient is being investigated for intermittent hematuria. Labs showed hemoglobin 8.1 with high reticulocyte count. Patient also complains of intermittent difficulty in swallowing and is on Warfarin therapy due to three episodes of portal vein thrombosis. What investigation you will order to confirm the probable diagnosis?

A. Bone marrow biopsy

B. ANA level

C. Cold agglutinin

D. [✓ Flow cytometry]

E. G6PD level

Flow cyto

Ans: Flow cytometry

Explanation: Anemia and high reticulocyte count with history of recurrent venous thrombosis with GI symptoms is classic paroxysmal nocturnal hemoglobinuria (PNH).The gold standard investigation is flow cytometry. Flow cytometric analysis using antibodies directed against GPI-AP is the most sensitive and informative assay available for diagnosis of PNH. PNH is a consequence of nonmalignant clonal expansion of one or several hematopoietic stem cells that have acquired a somatic mutation of PIGA. Progeny of affected stem cells are deficient in glycosyl phosphatidylinositol–anchored proteins (GPI-APs). Deficiency of the GPI-anchored complement regulatory proteins CD55 and CD59 accounts for the intravascular hemolysis that is the primary clinical manifestation of the disease. PNH frequently arises in association with disorders of bone marrow failure, particularly aplastic anemia. Thrombophilia is a major cause of morbidity and mortality in PNH. The classic manifestation of paroxysmal nocturnal hemoglobinuria (PNH) is dark urine during the night with partial clearing during the day. Hemolytic anemia is usually in the form of intravascular hemolysis. The most common presentation is the presence of anemia associated with dark cola-colored urine that is a manifestation of hemoglobinuria. Thrombosis involves the venous system, and it typically occurs in unusual veins, namely the hepatic, abdominal, cerebral, and sub dermal veins. Other symptoms of PNH include esophageal spasms that occur in the morning and, like the dark-colored urine, clear up later in the day. In males, erectile dysfunction can occur concomitantly with hemoglobinuria; the cause of this is unknown. The anticomplement agent eculizumab is a humanized monoclonal antibody against terminal protein C5; it has been shown to be highly effective in reducing intravascular hemolysis. Hematopoietic stem cell transplantation (HSCT) using allogeneic donors is the only curative therapy for PNH.

Cold
warm
&
gB

29. A 25 year old female presented with history of sudden onset of severe anemia over the 2 weeks. Patient also complains of joint pains and mouth ulcers. Examination showed pallor, jaundice and butterfly shaped skin rash on face. What investigation should be done for the cause of anemia?

A. Iron studies
B. Vitamin B12
C. [✓ Coombs test]
D. Bone marrow biopsy
E. Hb electrophoresis

Coo

Ans: Coombs test

Explanation: History and examination are suggestive of hemolytic anemia in a patient with SLE which is autoimmune hemolytic anemia. The diagnosis is done by Coombs test. Coombs test is an immunology laboratory procedure used to detect the presence of antibodies against circulating red blood cells in the body, which then induce hemolysis. It is also called antiglobulin test. Antiglobulin testing can be either direct antiglobulin testing (DAT) or indirect antiglobulin testing (IAT). The principle of DAT is to detect for the presence of antibodies attached directly to the RBCs, which takes place by washing a collected blood sample in saline to isolate the patient's RBCs; this procedure removes unbound antibodies that may otherwise confound the result. IAT, by contrast, is used to detect unbound antibodies to RBCs, which may be present in the patient's serum. Anemia is found in about 50% of patients, with anemia of chronic disease, being the most common form. Impaired erythropoietin response and presence of antibodies against erythropoietin may contribute to the pathogenesis of this type of anemia. Patients with autoimmune hemolytic anemia usually belong to a distinct category, which is associated with anticardiolipin antibodies, thrombosis, thrombocytopenia, and renal disease, often in the context of secondary antiphospholipid syndrome.

30. A patient with history of anemia and jaundice is investigated with a peripheral film which showed 3% fragmented RBCs (schistocytes). Which of the following is not a cause of this abnormality?
   - A. Hemolytic uremic syndrome ✓
   - B. Thrombotic thrombocytopenic purpara.
   - C. Mechanical heart valves
   - D. G6PD deficiency Heinz bodies Bite cells
   - E. Disseminated intravascular coagulation ,

Heinz bodies

Ans: G6PD deficiency

Explanation: Schistocytes are red blood cell fragments. The presence of schistocytes on a peripheral blood smear is a hematological emergency that requires prompt review and investigation for microangiopathic hemolytic anemia which includes HUS, TTP and DIC. Fragmentation of RBCs is produced by mechanical damage in the circulation and can also be seen in patients with mechanical heart valves or those receiving dialysis. The presence of ≥1% schistocytes on a PBS in the absence of other moderate RBC changes is an important criterion for the diagnosis of MAHA. G6PD deficiency causes Heinz bodies on peripheral film.

31. A patient presented with cough and fever for 3 days, chest x-ray showed bronchopneumonia. CBC showed low hemoglobin with high LDH and retic count. Patient also having skin rash with target lesion. Patient is started on azithromycin. What is the cause of anemia in this patient?
   - A. G6PD deficiency
   - B. Warm hemolytic anemia
   - C. Cold agglutinin disease ✓

WDUY

D. Alloimmune hemolytic anemia

E. Microangiopathic hemolytic anemia

Ans: Cold agglutinin disease

Explanation: This is a case of mycoplasma pneumonia. Acute cold agglutinin disease is commonly associated with mycoplasma or infectious mononucleosis. This occurs due to antibodies usually IgM which bind to the RBCs at low temperature and cause them to agglutinate. Cold antibodies bind best at 4C.

32. A 25 year old male presented with weakness and epistaxis. CBC showed Hb 7.1, WBC count of 1200 and platelets 30000. Reticulocyte are 1%. Peripheral film is normocytic normochromic. There is no hepatosplenomegaly or lymphadenopathy. What is your diagnosis?

A. Acute leukemia

B. Megaloblastic anemia

C. Hemolytic anemia

D. Myelofibrosis

E. Aplastic anemia ✓

Ans: Aplastic anemia

Explanation: Pancytopenia without lymphadenopathy and hepatosplenomegaly is aplastic anemia until proven otherwise. Camitta criteria for diagnosis of aplastic anemia is: to diagnose aplastic anemia there must be at least 2 of the following: Hb < 10, platelets < 50,000 and neutrophils < 1500.

33. A 35 year old patient has blood tests for anemia which showed Hb 9.4, platelets 15000, neutrophils 300 and Retic count 15000. Bone marrow aspiration showed hypo cellular marrow with 20% hematopoietic cells. What is your diagnosis?

3000

10, 50,000

740

A. Moderate aplastic anemia

B. Severe aplastic anemia

C. Very severe aplastic anemia

D. Mild aplastic anemia

E. Unclassifiable aplastic anemia

Ans: Severe Aplastic anemia

Explanation: The modified Camitta criteria is used to assess the severity of aplastic anemia as follow:

- **Severe AA**: marrow cellularity < 25% plus at least two of the following; neutrophils < 500, platelets < 20000 and Retic < 20000.
- **Very severe:** as for severe with neutrophils less than 200.
- **Non- severe AA**: not fulfilling criteria for severe or very severe Aplastic anemia.

34. What is the most common cause of death in aplastic anemia?

    A. Bleeding
    B. Heart failure due to anemia
    C. Iron overload
    D. Sepsis
    E. Kidney failure

Sporolina
Itq.

Ans: Sepsis

Explanation: Infections are the major cause of death in AA. Neutropenia in AA is prolonged and persistent resulting in a higher incidence of invasive fungal infection and severe bacterial sepsis. Severe neutropenic patients should be isolated and prophylactic antibiotics and itraconazole should be given.

35. A 45 year old male patient is diagnosed as a case of severe aplastic anemia which requires recurrent transfusions. Patient has no HLA matched donor available. What should be the next best management step for him?

    A. Horse ATG+ cyclosporine
    B. Rabbit ATG + cyclosporine
    C. Erythropoietin
    D. Mycophenolate + prednisone
    E. Unrelated stem cell transplant

30-50 ATG.

30-50 years

30-50

30-

K.

Ans: Horse ATG + cyclosporine

Explanation: The decision of treatment of AA is dependent on age of patient.

- **Age less than 35,** best treatment is HLA identical stem cell transplant if available, if not available then unrelated donor stem cell transplant, still not available, then horse ATG + cyclosporine.

- **For age 35- 50**, first line is HLA identical stem cell transplant, if not available then horse ATG+ cyclosporine. If no response after 3-6 months, unrelated donor stem cell transplant.
- **For age > 50**, horse ATG + cyclosporine is 1st line. If no response then stem cell transplant consider after evaluation.

There is no role of erythropoietin, mycophenolate or cyclophosphamide in the treatment of AA.

This patient comes in category 2 with age range of 35-50, so initial best management is identical bone marrow transplant which is not available. So the next best is horse ATG plus cyclosporine.

36. A 22 year old man with sickle cell disease requires a cholecystectomy for persistent abdominal pain secondary to cholelithiasis. His Hb is 7.5 with HbS 68%. His surgeon requests you to evaluate him preoperatively. What do you recommend?

    A. No treatment needed
    B. Exchange transfusion to an HbS< 30%
    C. Hydroxyurea and delay surgery
    D. Simple transfusion to Hb of 10
    E. Simple transfusion to Hb of 12

9·8

⑩ ⑦

Ans: Simple transfusion to Hb of 10

Explanation: Preoperatively, a conservative transfusion regimen to a goal of Hb 10 has less post-operative complications as aggressive transfusion with exchange transfusion to less than 30% HbS. In low to medium risk procedures, simple transfusion to Hb 10 is recommended in sickle cell patients.

37. A 43 year old man presents to the emergency with a 4 week history of worsening abdominal discomfort and weight gain. On physical examination, a plethoric complexion is noted. Abdomen is tense with fluid thrill. Lab findings are, Hb 18.9, WBC 9000, platelets 450,000, ALT of 84, AST of 107 and bilirubin 2.1. Doppler shows hepatic vein thrombosis. Which of the following test is most likely to establish the cause of patient presentation?

    A. CT of abdomen
    B. Flow cytometry for CD55 and 59
    C. JAK 2 mutation ✓
    D. Lupus anticoagulant
    E. Erythropoietin level

Ans: JAK 2 mutation

Explanation: This patient is having Budd Chiari syndrome due to thrombosis of hepatic veins. Patient is having high hemoglobin level which points towards polycythemia. Patients with polycythemia may experience thrombosis at unusual sites including the splanchnic and cerebral venous systems. Thrombosis of the hepatic veins can cause fulminant hepatic failure or present sub acutely with abdominal pain, weight gain and rapidly worsening ascites. The most common cause of Budd Chiari syndrome is an underlying myeloproliferative disorder most commonly polycythemia. Screening of JAK2 mutation is always recommended in patients presenting with BCS, regardless of the presence of clinical clues of myeloproliferative disorder. CT abdomen is used for liver carcinoma. Lupus anticoagulant is also not recommended in this case with no other manifestation. PNH can presents with BCS but it is associated with low hemoglobin and pancytopenia sometimes. Erythropoietin is used to differentiate primary and secondary polycythemia and is not a confirmatory test.

38. Which test is used to differentiate between true and spurious polycythemia?

A. Erythropoietin
B. Red cell mass ✓
C. Peripheral film
D. Hematocrit
E. Bone marrow aspiration

Retics

Red cells

Ans: Red cell mass

E

Explanation: Normal red cell mass with a low normal plasma volume suggests relative or spurious polycythemia. High red cell mass with normal plasma volume suggests true polycythemia. So red cell mass is used to differentiate between true polycythemia and relative polycythemia.

JAK 2 mutation or erythropoietin level are used to differentiate between primary and secondary polycythemia. (**If JAK 2 mutation and erythropoietin both are given in options, then mark JAK 2 mutation).**

39. Which of the following is not a feature of polycythemia rubra vera?

A. Hyperurecemia ✓
B. Splenomegaly ✓
C. Raised iron stores in BM ✓
D. Risk of thrombosis ✓

LDH

Pcv

plasma

RJ

E. Basophilia

Ans: Raised iron stores in BM

Explanation: Iron stores are typically absent in BM in patients with polycythemia vera, having been transferred to the increased circulating red blood cell mass. There may be concomitant iron deficiency resulting from chronic gastrointestinal blood loss due to high incidence of PUD in these patients.

PRV is associated with hyperurecemia due to overproduction of uric acid. Spleen is clinically palpable in 75% of patients and nearly 100% enlarged when imaged by USG. Thrombosis is the most common complication of PRV, due to increased blood viscosity and abnormal platelet functions, is the major cause of morbidity and mortality. Basophilia and eosinophilia are frequently present in PRV.

40. A 54 year old male is recently diagnosed as JAK2 mutation positive PRV. He complains of itching and headache. CBC showed Hb 20, HCT 61%, WBC of 13000, and platelets of 690000. What is the next best management for him?

JAK-2

A. Low dose aspirin

B. High dose aspirin

C. Low dose aspirin+ venesection with HCT<45% ✓

D. Low dose aspirin+ venesection with HCT 45-50%

E. Aspirin + Hydroxyurea

Rux

Ans: Low dose aspirin + venesection with HCT < 45%

Explanation: People younger than 60 years and no history of thrombosis are labeled as **low risk PRV**. They should receive low dose aspirin to reduce the risk of thrombosis and control vasomotor symptoms. High dose aspirin has risk of bleeding. Patient should also receive venesection to achieve and maintain a hematocrit below 45%. Hydroxyurea is reserved for high risk patients and for low risk patients who do not tolerate venesection or require frequent venesection or who have disease related bleeding, severe symptoms, symptomatic splenomegaly or progressive leukocytosis. Patient whose disease is progressive after treatment with hydroxyurea, ruxolitinib (Jakafi) is next treatment.

41. A 42 year old woman who has AML is stable and having no fever and no bleeding. She will get chemotherapy after 2 weeks. At what platelets counts, she should receive a platelet transfusion?

A. 3000

B. 10000

C. 20000

D. 30000

E. 50000

Ans: 10000

Explanation: Patient with no bleeding => transfusion at 10000, patient with bleeding => platelets transfusion at 50000. In pregnancy, start transfusion below 20000 with no bleeding.

42. A patient was admitted with an upper GI bleed received a red cell transfusion. 1 hour after the transfusion, he complains of severe dyspnea, over the next 30 minutes he develops respiratory failure and fever with a chest x ray showing infiltrates. He is intubated and over the next 48 hours extubated successfully. What is the probable cause?

A. Acute hemolytic reaction

B. Allergic reaction

C. Transfusion related infection

D. Graft vs. host disease

E. Transfusion related acute lung injury

10'

50,000

10P. 30

9.8

Ans: Transfusion related acute lung injury

Explanation: The scenario of rapid onset of respiratory failure with transfusion that rapidly resolves over a short period of time is most consistent with transfusion related acute lung injury. Hemolytic reaction presents with fever and low back pain with hemoglobinuria. GVHD involves many organs but not lungs. Transfusion related infection are rare and they are not settled within 48 hours.

43. Fresh frozen plasma is indicated in all of the following except:

A. Severe factor V deficiency and upcoming surgery

B. TTP initial therapy

C. Cirrhosis with INR 1.9

D. Massive blood transfusion

E. Warfarin induced bleeding

FFP

Ans: Cirrhosis with INR 1.9

Explanation: Patients with cirrhosis and mild INR elevation without bleeding do not respond to plasma and unlikely to have significant coagulation defects. Patients with documented factor deficiency, warfarin induced bleeding, and TTP benefit from FFP transfusion. Patient who needs massive transfusion should also get 1:2 FFP transfusion to prevent massive transfusion related coagulopathy.

44. A call is sent from surgical emergency for a polytrauma patient with history of bleeding from the venipuncture site. Patient has been given 14 units of PRBCs in the last 48 hours along with calcium IV. Labs showed HB 10.2, platelets 170000, INR 3.2, PTT 60 seconds with normal fibrinogen. What is the most appropriate therapy for him?

    A. Cryoprecipitate
    B. Desmopression
    C. Fresh frozen plasma ✓
    D. Topical thrombin
    E. No treatment

AML
10,000

aptt ↑

RBCs
VIII, XIII, VWF

proincomb
FFP
aptt

Ans: Fresh frozen plasma

Explanation: Following massive blood transfusions, dilutional coagulopathy develops and is best treated with fresh frozen plasma containing all coagulation factors. Cryoprecipitate is rich in factors VIII, XIII, vW factor and fibrinogen and is used when fibrinogen is less than 100. Topical thrombin is only suitable for brief period. Desmopression is used in vWD.

IV.00 9.8

45. A diagnosed case of IgA deficiency patient with celiac disease needs blood transfusion before surgery. Which of the following should be recommended in this patient for transfusion?

    A. Diphenhydramine before transfusion
    B. Hydrocortisone before transfusion
    C. Leukoreduction
    D. Washed RBCs
    E. Irradiated RBCs

30000
10000

1.7

jiyan
B. opry

1,000
2000

Ans: Washed RBCs

Explanation: IgA deficient patients have antibodies against IgA. Interaction of these antibodies with donor IgA will cause transfusion reaction. Washing of RBCs reduces IgA in the donor. Leukoreduction does not reduce IgA contents. Steroids and antihistamine have no role in prevention of this reaction.

46. Which of the following blood products have highest risk of bacterial contamination?

    A. Whole blood

B. PRBCs

C. Platelets

D. FFPs

E. Cryoprecipitate

Ans: Platelets

Explanation: Platelets are more prone to bacterial contamination because they cannot be refrigerated. Bacterial contamination occurs in 1:30000 PRBC donation and 1:5000 of platelets donations.

47. You are transfusing platelets to one of patient with ITP before surgery. Your house officer asked you that after transfusing one pack of platelets, how much platelets will be increased?

A. 15-20000

B. 20-25000

C. 5-10000 ✓

D. 30-40000

E. 40-50000

5

30,000

50,

Ans: 5-10000

Explanation: A platelet concentrate single bag containing approximately 0.7 X 1011 platelets should cause a platelet count increase of 5000 to 10,000 / µL in an average sized adult. 6 bags from multiple donors or one megaconcentrate through apheresis is commonly transfused in a patient who needs platelets transfusion which will raise platelets approximately 30,000 to 60,000.

48. A 26 year old female has developed urticaria during blood transfusion. She is otherwise normal. There is no fever or dyspnea. Transfusion stopped and dimenhydrinate given. Which component of blood causes such type of reaction?

A. RBCs

B. Donor Leucocytes

C. Plasma proteins ✓

D. Recipient leucocytes -

E. Antibodies of donor

Pro

Ce

VII XIII VF!

procyclidine

clozapine

T

Ans: Plasma proteins

Explanation: Hypersensitivity reactions causing urticaria or bronchospasm are almost always due to exposure to plasma proteins. Leukoagglutination reactions occur due to antigens on leucocytes. Mismatched RBCs cause hemolytic transfusion reactions. Donor Leucocytes causes GVHD in high risk patients.

49. A patient with a diagnosis of AML on chemotherapy needs whole blood transfusion. Patient is severely neutropenic. What precaution should be done to reduce the risk of transfusion related graft vs. host disease?
    - A. Leucoreduced blood
    - B. Washed RBCs A
    - C. Irradiated blood
    - D. Leukopoor blood
    - E. Frozen Blood

Ans: Irradiated blood

Explanation: TA-GVHD occurs when there is unrestrained proliferation of transfused donor lymphocytes. This occurs when the recipient's ability to mediate elimination of the donor cells following transfusion is compromised either because of the lack of genetic disparity between the donor and recipient or because the recipient's immune system is suppressed. Bone marrow aplasia induced by donor anti-recipient responses makes it difficult to reverse the course of TA-GVHD once it has begun. Thus, the focus for the prevention of TA-GVHD has been to prevent donor T cell proliferation. Currently, γ-irradiation is used to prevent TA-GVHD. So any patient who is severely immunocompromised should get irritated blood products.

PL - Prothwmy

50. A patient who is a diagnosed case of AML-M3 presented with epistaxis. CBC showed Hb 11.2, WBC 20000, platelets 60,000, PT 24 seconds, APTT 60 seconds and fibrinogen 85. What should be the next treatment step?
    - A. Antibiotics
    - B. Platelet transfusion
    - C. FFP transfusion ✓
    - D. Cryoprecipitate transfusion
    - E. Whole blood transfusion

VIII / XIII : VWF

1 gld

VIII

XII

VWF

Fib

Ans: Cryoprecipitate transfusion

Explanation: This is a case of tumor associated DIC which is commonly associated with AML M3 (APML). The most important clinical feature of APML is life-threatening hemorrhage, which is caused mainly by enhanced fibrinolytic-type disseminated intravascular coagulation. Laboratory findings of DIC in patients with APML are characterized by marked hypofibrinogenemia and an increase in ratio of fibrin/fibrinogen degradation products (FDP)/D-dimer. Initial treatment should be done with cryoprecipitate to keep fibrinogen level more than 150. FFPs are used when cryoprecipitate not available or when PT, APTT are high with normal fibrinogen. Platelets should be kept above 50000. Patient should be given all trans retinoic acid (tretinoin, Vesanoid) therapy. Antibiotics has no role in cancer related DIC.

51. A 62 year old patient presented with complaints of burning of the hands associated with redness in both hands. CBC done which showed platelets of 100, 0000 with normal hemoglobin and WBC. Peripheral film shows large platelets. What is the treatment for this patient?
    - A. Aspirin+ hydroxyurea ✓
    - B. Aspirin + anagrelide
    - C. Hydroxyurea
    - D. Aspirin
    - E. Peg interferon alpha 2

Ans: Aspirin+ Hydroxyurea

Explanation: This patient is having essential thrombocytosis. These patients have high risk of thrombosis at unusual sites so platelets should be kept below 500000. The treatment of choice is hydroxyurea plus low dose aspirin. Anagrelide is used when hydroxyurea is not well tolerated.

Bleeding with ET => plateletpheresis. Bleeding in ET occurs due to acquired vWD associated with very high platelets count.

52. A 55 year old presented with left sided abdominal pain. Examination showed massive splenomegaly. CBC showed pancytopenia. Peripheral film showed poikilocytosis with teardrop cells, large degranulated platelets and promyelocytes. What is your diagnosis?
    - A. Severe infection
    - B. Metastatic carcinoma to marrow
    - C. Primary myelofibrosis
    - D. Chronic myelocytic leukemia
    - E. Myelodysplasia ✓

Ans: Primary myelofibrosis

Explanation: The tetrad of massive splenomegaly, teardrop cells, leukoerythroblastic blood and giant abnormal platelets are typical of myelofibrosis. Myelofibrosis is also associated with dry tap on bone marrow aspiration in advanced cases.

53. A patient with pancytopenia has a bone marrow biopsy which shows hyper cellular marrow. Which of the following is less likely the cause of this presentation?

A. Megaloblastic anemia
B. SLE
C. Hypersplenism
[✓ D. Hairy cell leukemia]
E. Myelodysplasia

Ans: Hairy cell leukemia

Explanation: Hairy cell leukemia produces dry tap. All the other causes are associated with hyper cellular bone marrow with pancytopancytopenia.

Pancytopenia with hypocellular marrow:

- Acquired aplastic anemia
- Inherited aplastic anemia
- Some myelodysplastic syndromes
- Myelofibrosis
- Overwhelming infections
- Toxic depression of BM
- Anorexia nervosa
- PNH

Pancytopenia with cellular marrow:

- Myelodysplasia
- Lymphoma
- Leukemia
- Myeloma

- SLE
- Hypersplenism
- Vitamin B12/ folate deficiency
- Alcoholism
- Disseminated TB
- Sarcoidosis
- HIV

54. A patient with platelets of 100, 0000 with Hb of 14.1. What investigation should be done in this patient?

    A. JAK 2 mutation

    B. BCR-ABL gene

    C. Karyotyping

    D. CT of chest and abdomen

    E. Iron studies

600×10^9

VIII XIII

VII

Ans: JAK 2 mutation

Explanation: A JAK 2 mutation is present in 40-50% of patients with essential thrombocytosis and should be done in all patients with the diagnosis of ET. Iron studies should be done for reactive thrombocytosis in moderate to severe anemia. Reactive thrombocytosis rarely has platelets more than 600,000.

55. A patient presented with weakness and petechiae on lower limbs for 1 week. CBC showed Hb 8.3, WBC 10,000 and platelets 30,000. Reticulocyte count is 10% with high LDH level. What is your diagnosis?

    A. Vitamin B12 deficiency

    B. Aplastic anemia

    C. ITP ✓

    D. Evan syndrome ✓

    E. Autoimmune hemolytic anemia

ITP

Ans: Evans syndrome

Explanation: Vitamin B12 can cause low Hb and platelets but it is associated with low retic count. Aplastic anemia is also associated with low retic count. ITP can be associated with anemia due to bleeding and can cause high retic but LDH will be normal. Autoimmune hemolytic anemia alone does not cause low platelets. So most likely diagnosis is Evans syndrome which is a combination of autoimmune hemolytic anemia and ITP.

56. A 65 year old male presented with fatigue and left upper quadrant discomfort. Examination showed pallor with splenomegaly. CBC showed Hb 10, WBC of 80,000 with no peripheral blasts and normal platelets. Peripheral film showed myelocytes and metamyelocytes with high basophils. What cytogenetic changes should be seen to confirm your diagnosis?

- A. T (8; 21)
- B. Inv (16)
- [✓ C. T (9; 22)]
- D. Del (5)
- E. t (14;18)

LDH

Ans: t (9; 22)

Explanation: t (9; 22) also called Philadelphia chromosome is the defining feature of CML. It results in fusion gene BCR-ABL which leads to tyrosine kinase activity and ultimately clinical phenotype of CML. t (8; 11), and inv (16) are present in AML. Del (5) is most commonly seen in myelodysplasia.

57. What is the first line treatment for CML?

- A. Interferon
- B. Hydroxyurea
- [✓ C. Imatinib]
- D. Nilotinib
- E. Interferon plus cytarabine

Ans: Imatinib is the treatment of first choice as first line treatment for CML. Nilotinib is second line drug.

58. Which tyrosine kinase inhibitor is associated with the development of pleural effusion?

- A. Bosutinib
- [✓ B. Dasatinib]
- C. Imatinib
- D. Nilotinib

E. Ponatinib

Ans: Dasatinib

Explanation: Dasatinib is associated with development of pleural effusion in 28% of treated patients.

59. A 26 year old female presented with bruising and fatigue. CBC showed Hb 8.4, WBC 60,000 with 60% blasts and platelets 40,000. Peripheral film showed blast cells with Auer rods. What is the diagnosis?

    A. ALL
    B. CLL
    C. CML
    D. AML ✓
    E. Myelodysplasia

plasma

760. years

120 years

5, 7

Ans: AML

Explanation: Marrow failure with high blasts cells and presence of Auer rods are diagnostic of AML.

60. A 24 year old man presented with dyspnea, fatigue and headache for 3 days. Labs showed WBC of 143,000 with 70% blasts, hemoglobin 7.7, platelets 35000, creatinine 2.1, K 5.7, Calcium 6.6, P 7.3 and uric acid 13.2. What is the next best management step for him?

    A. IV fluids and allopurinol
    B. Immediate chemotherapy
    C. RBCs transfusion
    D. Rasburicase, allopurinol and leukopheresis ✓
    E. Platelets transfusion

Hyperkal

FCR

CR

Ans: Rasburicase, allopurinol and leukopheresis

Explanation: This patient has acute leukemia with hyper leukocytosis which is defined as WBC more than 100,000. There are three main complications of hyper leukocytosis: DIC, tumor lysis syndrome and leukostasis. This patient has tumor lysis syndrome which is defined as 2 or more abnormal laboratory values of uric acid, K, PO4 or calcium. Prevention of TLS is done with hydration and allopurinol. Treatment is with hydration, allopurinol and Rasburicase. Leukopheresis should be done prophylactically to reduce the risk of leukostasis.

61. A patient with lymphadenopathy and splenomegaly has CBC with peripheral film which showed low hemoglobin, low platelets and WBC of 25000. On peripheral film, WBCs are high in number with pale blue cytoplasm, prominent nucleoli and hair like projections. What is the diagnosis?

A. ALL
B. AML
C. CML
D. [✓ Hairy cell leukemia]
E. Non-Hodgkin's lymphoma

WBCs ↑

CR

Ans: Hairy cell leukemia

Explanation: Marrow failure with splenomegaly and lymphadenopathy with blue cytoplasm and hair like projections are suggestive of hairy cell leukemia. Bone marrow biopsy should be done for complete evaluation. Cladribine is treatment of choice.

62. Which acquired genetic alteration is nearly universally present in classical Hodgkin lymphoma?

A. t(14;18)
B. [✓ 9p24.1 amplification]
C. 17 p deletion
D. C-MYC translocation
E. Inv (16)

C3-Cu

clue 9p24:1

Ans: 9p24.1 amplification

Explanation: Genetic alteration in the 9p24.1 is universally present in classical Hodgkin's lymphoma. It is defining feature of cHL.

lytic

63. A 65 year old patient has low back pain not getting better with treatment with NSAIDs. A MRI of spine showed a T2 enhancing lesion in the lumber vertebra, bone scan is done which came out to be negative for Mets. What laboratory test would most likely to be positive in this case?

A. Beta HCG
B. PSA level
C. [✓ Serum protein electrophoresis]
D. CEA level
E. CA 19-9

lytic lesion

Ans: Serum protein electrophoresis

Explanation: Progressive back pain with enhancing lesion on MRI with negative bone scan indicates that patient has purely lytic lesion. Bone metastasis are usually sclerotic or blastic. Only four cancers can cause lytic bone lesions; lung, thyroid, kidney and breast. The most common cause of lytic bone lesion is multiple myeloma due to inhibition of osteoblastogenesis and promotion of osteoclastic activity. A serum protein electrophoresis test should be done which is positive in more than 95% of patients. CEA is raised in bowel cancer, PSA in prostate cancer, CA 19-9 in pancreatic cancer and HCG is raised in germ cell tumors which all produce sclerotic lesions.

Bones

64. Which one of the following parameters is not used for the staging of multiple myeloma?

A. Albumin

B. LDH level

C. Beta-2 microglobulin

D. Serum calcium

E. [✓ Tumor cytogenetic]

Ans: Serum calcium

MRI

Explanation: The revised International Staging System use albumin, beta 2 microglobulin, LDH level and tumor cytogenetics for staging of MM at diagnosis. There is no role of hemoglobin and calcium in staging.

65. Which of the following clinical signs is not a defining characteristic of multiple myeloma that requires treatment?

A. Anemia ✓

B. [✓ Hematuria]

C. Hypercalcemia ✓

D. Lytic bone lesions ✓

E. Renal insufficiency ✓

CR AB

CRAB

Ans: Hematuria

Explanation: Hematuria is not usually a sign of MM at diagnosis. Common symptoms are CRAB; hyperCalcemia, Renal insufficiency, Anemia and lytic Bone lesions. Frequent infections due to immunoparesis is also a feature of MM.

66. A 75 year old female came for investigation of an elevated total protein level found on routine testing. Patient is otherwise asymptomatic. Further testing shows normal Hb and Calcium level. Skeletal survey is also normal. 24 hour urinary protein is also normal. Bone marrow biopsy showed 6% plasma cells. What is your diagnosis?

30% MM.

A. Light chain deposition disease

B. Monoclonal gammopathy of undetermined significance

C. Asymptomatic MM

D. Smoldering MM

E. Walderstorm macroglobulinemia

MGUS

5 30 50

1 70 100

Ans: Monoclonal gammopathy of undetermined significance

30%

Explanation: MGUS is defined as bone marrow plasma cells less than 10%, paraprotein less than 3 and lack of typical CRAB symptoms of MM. Light chain disease is ruled out by the normal 24 hour urinary protein. Smoldering MM has plasma cells more than 10% but less than 20%.

67. A diagnosed case of MM presented with sudden onset of lower limb weakness associated with urinary incontinence and severe back pain for 5 hours. MRI of spine showed mass lesion at L4, L5 vertebra. What is the next best course of action?

mets

A. High dose dexamethasone + radiation

VIII Dexa.

B. High dose dexamethasone + urgent decompression

C. Start chemotherapy

D. Dexamethasone + Zolendronic acid

E. High dose dexamethasone and observe

Ans: High dose dexamethasone + radiation

Explanation: This patient has presented with acute spinal cord compression due to MM which is a neurological emergency which must be treated with urgent decompression. Both surgical decompression and radiation therapy are used to treat compression. The method is determined by the sensitivity of tumor cells to radiation. Because myeloma cells are highly radiosensitive, so this is the method of choice for new cord compression. High dose dexamethasone is required as well to help reduce the size of the mass as well as mitigate any potential edema that may result from the initial sessions of radiation. All other treatment are used after radiation.

68. Which of the following tests or combination of tests are used for prognosis of MM?

A. Albumin

B. Beta 2 microglobulin

C. Albumin + calcium

D. Albumin + beta 2 microglobulin ✓

E. Beta 2 Microglobulin + calcium

Ans: Albumin and beta 2 microglobulin are used to determine the progno sis of the MM.

69. A 34 year old female diagnosed case of ITP has steroid dependent di'sease. Which of the following is the best option for her for achieving long term remission?

A. Eltrombopag

B. Rituximab 4 weekly

[✓ C. Splenectomy] ✓

D. IVIG monthly

E. Romiplostim

IVIG In 6 hours
Stso 3-4 days
10,000

Ans: Splenectomy

Explanation: The indications for splenectomy in ITP includes:

- Failure to respond to steroids.
- Inability to taper steroids or steroids dependent ITP
- recurrent ITP

Vw XTV
50,000
80,000

Splenectomy is associated with an immediate response rate of 80% to 90% and a long-term response rate of 60% to 70%. Long term remission is much better than with rituximab therapy. IVIG are excellent therapy for acutely rising the platelets counts before an emergency surgery. Eltrombopag and Romiplostim are thrombopoietin agonists have high response rates but platelets counts return to baseline when stopped.

70. A 25 year old female presented with easy bruisbility for 3 months. She is otherwise asymptomatic with no history of bleeding. CBC showed normal Hb with platelets of 40,000. There is no hepatosplenomegaly or lymphadenopathy. What should be done? 4

A. Anti- D immunoglobulin

B. IVIG therapy

C. Observation

D. Prednisone to increase platelets above 50000

E. Rituximab

Ans: Observation

Explanation: This patient is having ITP, 1st line treatment is steroid but steroid should be given when the platelet count is less than 30,000. The best course of action is observation at this stage with monthly CBC result.

30,000

71. Which of the following findings would be most helpful in making a diagnosis of ITP most likely?
    - A. [✓ Increased megakaryocytes on bone marrow]
    - B. Elevated reticulated platelets
    - C. [✓ Response to steroids]
    - D. Antiplatelet antibodies
    - E. Normal thrombopoietin level

IVIG

bone → (2) biopsy

Ans: Response to steroids

Explanation: The patient response to steroids is very specific for ITP as most primary marrow disorders causing low platelets would not respond to steroids. The presence of antiplatelet antibodies is neither specific nor sensitivite for ITP. The presence of slightly increased megakaryocytes on bone marrow is also nonspecific for ITP, bone marrow exam is mostly used to exclude amegakaryocytic thrombocytopenia.

72. A 15 year old boy presented with fever, bruising and fatigue for 10 days. CBC showed Hb 7.3, WBC 60,000 with 70% blasts cells and platelets of 60,000. Bone marrow biopsy showed hyper cellular marrow with 80% blasts which stains positive for tdt. CXR showed mediastinal mass. What is the diagnosis?
    - A. [✓ ALL]
    - B. AML
    - C. Large cell lymphoma
    - D. Hodgkin's lymphoma
    - E. Burkitt lymphoma

td.

ALL.

Ans: ALL

Explanation: Marrow failure with 80% blasts which are tdt positive is typical of ALL. In AML, blasts cells contain Auer rods. In ALL, patient can present with SOB due to mediastinal mass.

73. A patient with a history of ITP admitted with history of intestinal obstruction which needs laparotomy, surgeon wants you to assess the patient regarding management of platelets. What target you will advise to prevent excessive bleeding?

- A. > 30,000
- B. > 50,000 to 80,000
- [✓ C. > 80,000-100,000]
- D. > 20,000
- E. > 150,000

5

Ans: > 80,000 -100,000

Explanation: For minor surgery => > 50,000-80,000

For major surgery => >80,000-100,000

5-10 HIT

74. A patient was admitted for treatment of NSTEMI 1 week back which was managed with antiplatelets and unfractionated heparin. Patient was discharged successfully 2 days back. Today, patient presented with complaints of right leg swelling. CBC done which showed platelets 25,000 which were 250,000 at initial admission. Doppler USG leg showed proximal DVT. What should be your next management step?

- A. LMWH
- B. Warfarin
- [✓ C. Argatroban]
- D. Tirofiban
- E. Platelets transfusion

NSTEMI

HIT

HIT

LMWH

AF

50%

[illegible]

Ans: Argatroban

Explanation: This is a case of heparin induced thrombocytopenia based on clinical picture of low platelets after 1 week of heparin therapy with DVT. A presumptive diagnosis of HIT is made when new onset thrombocytopenia is detected in a patient within 5 to 14 days of exposure to heparin. It should be confirmed with PF4-heparin antibodies test. Management of HIT involves;

Observe

- Discontinuation of all forms of heparin
- Begin treatment with direct thrombin inhibitor, the drug of choice is Argatroban.
- Perform Doppler ultrasound of lower extremities to rule out DVT.

- Follow platelets count daily until recovery occurs
- Transition anticoagulation to warfarin when platelet count has recovered, treat for 3 to 6 months in which thrombosis has occurred and for 30 days when thrombosis not occurred.

75. Which monoclonal antibody is most commonly present in Multiple myeloma?

A. IgG ✓

B. IgM

C. IgA

D. IgD

E. Light chains

30,000 pyr

~~Steroids~~ Azathio

IVIG.

Typo-

Ans: IgG

Explanation: Overall, the paraprotein in MM is IgG 60%, IgA 20%, light chain only 15% and remaining 5% is formed by IgM and IgD.

76. Which of the following combination of electrolyte abnormalities would suggest tumor lysis syndrome?

A. High K, high Ca, low P

B. Low K, high Ca, low P

C. High K, low Ca, high P ✓

D. Low K, low Ca, high P

E. Low K, low Ca, low P

10,000

Ans: High K, low Ca, high P

Explanation: Tumor lysis syndrome is a potentially deadly condition related to the treatment of high grade lymphomas and leukemias. Tumor lysis syndrome occurs from the breakdown of the tumor cells and the subsequent release of chemicals from the cells. It leads to a high potassium and high phosphate level in the presence of a low calcium. It could also cause an acute kidney injury in the presence of a high phosphate and high uric acid level. Hydration and allopurinol is used for prevention of TLS in low risk patients and hydration with Rasburicase is used in high risk patients. Aggressive hydration with saline plus Rasburicase and calcium gluconate are used for treatment.

77. A 62 year old patient is referred to you from ophthalmologist with history of headache and blurring of vision. Fundoscopy showed dilated and tortuous veins. Examination showed pallor

with axillary lymphadenopathy and hepatosplenomegaly. Labs showed low hemoglobin with high LDH. Serum electrophoresis showed IgM paraprotein band at 6.5g/l. A bone marrow biopsy showed 18% lymphoplasmacytic cells. What is your diagnosis?

A. Multiple myeloma

B. Monoclonal gammopathy of undetermined significance

C. Chronic lymphocytic leukemia

D. Waldenstorm macroglobulinemia ✓

E. Smoldering myeloma

Ans: Waldenstorm macroglobulinemia

Explanation: There is an enormous IgM paraprotein band on serum electrophoresis suggesting differential of multiple myeloma, Waldenstorm macroglobulinemia, and MGUS. The patient is symptomatic, has bone marrow involvement of greater than 10% with IgM band greater than 3g ruling out MGUS. There is lack of bone symptoms and renal involvement and paraprotein is IgM which is less likely to be present in MM. Also patient has symptoms of hyper viscosity which are also commonly present in Waldenstorm macroglobulinemia. Tortuous and dilated veins are classical in hyper viscosity with Waldenstorm macroglobulinemia. R ←

78. A 55 year old diabetic male presented with high grade fever with cough and SOB for 5 days. Labs showed Hb 10.3, WBC 20,000, platelets 25000, PT 25 second, PTT 60 seconds. There is no history of bleeding. What is the next best step in management of this patient?

A. FFP transfusion ✓

B. Whole blood

C. Platelets transfusion

D. Broad spectrum antibiotics

E. Cryoprecipitate transfusion

VIII XIIII

DIC

VIIII X-ray

aptt

Ans: Broad spectrum antibiotics

Explanation: This diabetic patient is having Sepsis related DIC with low platelets and high PT and APTT. The cornerstone DIC treatment is the treatment of the underlying cause which is sepsis due to pneumonia in this case, so broad spectrum antibiotics is the best next step. DIC spontaneously resolved in many cases when the underlying disorder is properly managed and improved. The administration of platelet concentrate, fresh frozen plasma or whole blood in patients with DIC is used with active bleeding or those at higher risk of bleeding requiring invasive procedures. Cryoprecipitate is used when fibrinogen is less than 100 and patient is bleeding.

79. Which one of the following test is more specific for the diagnosis of DIC?
    - A. Platelets
    - [✓ B. Elevated D- dimers]
    - C. Elevated PT
    - [✓ D. Fibrinogen level]
    - E. APTT

Ans: Elevated D- dimers

Explanation: Although combination of all these tests is used for the diagnosis of DIC and no single test is used for diagnosis, but D- dimers and FDPs are the most specific tests for DIC. D-dimers test is better than all and better than FDPs. All these tests are used in the scoring system of DIC for diagnosis. The negative predictive valve of D- dimer is used to exclude DIC.

80. Which of the following test is used to check the severity of DIC?
    - A. Low platelets
    - B. High PT
    - C. High PTT
    - D. High D- dimers
    - [✓ E. Low fibrinogen]

Ans: Low fibrinogen

Explanation: Fibrinogen is used to access severity of DIC. In most severe DIC, fibrinogen is mostly less than 100.

0

81. Which antibiotic is associated with the development of DIC in trauma patients?
    - A. Amoxicillin
    - [✓ B. Cefotetan]
    - C. Azithromycin
    - D. Levofloxacin
    - [✓ E. Co- trimaxazole]

pen Cipro

Ans: Cefotetan ✓

Explanation: A rare but fulminant form of DIC is seen with second and third generation cephalosporins specially Cefotetan. Hallmarks are massive immune hemolysis, severe DIC and thrombosis. The timing is within 2 weeks after receiving the drug. Most often occurs in trauma patients or after using for surgical prophylaxis.

82. A 28 year old female who is 10 days post-partum after spontaneous vaginal delivery presented with worsening confusion over the last 3 days. Examination showed pallor, multiple bruises over the arms with feverish feeling. CBC showed low hemoglobin, high WBC with low platelets, PT and APTT are normal. Patient is moving all four limbs. What is the most likely diagnosis?

   A. Hemolytic uremic syndrome
   B. Cavernous sinus thrombosis
   C. Thrombotic thrombocytopenic purpara ✓
   D. Disseminated intravascular coagulation
   E. Encephalitis

Ans: Thrombotic thrombocytopenic purpara

Explanation: TTP is CPSP favorite topic. The history of new onset confusion after delivery has few important differentials: TTP, HELLP, cerebral venous thrombosis, encephalitis, and intracranial bleeding. TTP is suggested by fever, hemolysis, low platelets, and kidney injury. In HUS, fever is less likely and confusion is minimal or none, CVT presents with sudden confusion which may also cause focal neurological deficit. It can also presents with seizures. Encephalitis is not associated with low Hb and platelets.

In this patient, the most probable cause is TTP, pregnancy and post-partum state accounts for 20% of cases. Severe deficiency of ADAMTS13 is the underlying cause for developing TTP. In acquired TTP this is mediated by auto-antibodies against ADAMTS13. An ADAMTS13 value of < 10% is indicative of TTP. However, ADAMTS13 levels can also be lowered (but > 10%) in pregnancy, postpartum, and other non-TTP diseases. The presence of schistocytes, the fragmented RBCs injured by damaged endothelium, in blood film investigation is one of the strong indicators of hemolytic anemia. A threshold of 0.2–0.5% schistocytes is necessary before suspecting TTP.

83. A patient is started on clopidogrel 2 weeks back for STEMI presented with confusion and fever. Labs showed Hb 10.3, platelets 50,000, and LDH 3000. PT, APTT are normal. Peripheral film showed schistocytes. What should be the next best management step?

A. Whole blood transfusion

B. Platelets transfusion

C. [✓ Plasma exchange]

D. Antibiotics

E. Steroids

Ans: Plasma exchange

Explanation: This is a case of TTP. Clopidogrel is rarely associated with development of TTP. Immediate administration of plasma exchange is treatment of choice as mortality is more than 95% without treatment. Plasma exchange is done once daily in TTP. Blood transfusion and platelets transfusion should be avoided in TTP because they cause propagation of platelet rich micro thrombi. Steroids alone has no role in TTP management but they can be used along with plasma exchange in relapse cases.

84. A known case of SLE patient is diagnosed as having TTP. Plasma exchange is not available urgently. In the meanwhile what treatment you will recommend?

A. Blood transfusion

B. [✓ FFP transfusion]

C. Platelets transfusion

D. Hemodialysis

E. Steroids

Ca VIII XIII

FFP.

VIII XIII

XII

Ans: FFP transfusion

Explanation: If plasma exchange is not available immediately, FFP can be given initially.

85. The patient of TTP is started on once daily therapeutic plasma exchange. How will you monitor the response to treatment?

A. Hemoglobin+ Platelets

V/S

B. Hemoglobin+ LDH

C. [✓ Platelets+ LDH]

D. LDH+ Bilirubin

E. LDH+ reticulocyte count

VIII XII

cryo

VIII XII

VWF Fib

Ans: LDH+ Platelets

Explanation: LDH and platelets count are used for monitoring the response to the plasma exchange, once the platelets and LDH return to normal for 2 consecutive days, the treatment is tapered off slowly with monitoring for the relapse.

86. Patient of TTP was managed successfully initially and plasma exchange has been stopped. Patient has normal platelets and LDH and no schistocytes were present on PF. 2 months after treatment, patient again develops TTP. What should be the best treatment now?
   - A. Plasma exchange
   - B. Plasma exchange+ rituximab ✓
   - C. Plasma exchange+ Steroids
   - D. FFPs
   - E. Splenectomy

Ans: Plasma exchange+ rituximab

Explanation: Rituximab is used for the treatment of relapse of TTP along with plasma exchange and for the prevention of relapse in relapsing disease. Methylprednisolone can also be used if rituximab is not available. Splenectomy is used for relapsing disease if rituximab is not available or contraindicated. There is no role of splenectomy in treatment.

87. Which of the following suggests TTP?
   - A. Creatinine 6.9
   - B. Associated with shiga toxin
   - C. ADAMTS 13 level 7% ✓
   - D. Raised PT, APTT
   - E. Increase FDPs

30'

AL

[illegible]

AA

Ans: ADAMTS 13 level 7%

Explanation: ADAMTS 13 level < 10% suggests TTP. Although kidney injury is a feature of TTP, but creatinine rarely raised more than 2.0. Marked kidney damage is a feature of HUS. HUS is also associated with Shiga like toxin. PT, APTT are typically normal in TTP and FDPs are slightly increased.

88. Classification of von Willebrand disease is based on which of the following?
   - A. Clinical phenotype and pathophysiological mechanisms
   - B. Factor VIII level

C. Genetic mutation

D. Platelets level

E. Bleeding time

Ans: Clinical phenotype and pathophysiological mechanisms

Explanation: The international society of thrombosis and hemostasis classification of von Willebrand disease incorporates important aspects of clinical phenotype, pathophysiological mechanisms and treatment considerations. The three types are;

- Type 1, which is a partial quantitative deficiency
- Type 2, which is a qualitative defect. It has 4 subtypes
- 2b Type 3, total absence of VWF.

89. Thrombocytopenia is associated with which type of von Willebrand disease?

A. Type 1

B. Type 2A

C. Type 2B

D. Type 2M

E. Type 3

2b [illegible] 2 [illegible] PTH

2b

Ans: Thrombocytopenia is associated with type 2B vWD.

Absence

90. Which of the following is not a treatment option for type 2B vWD?

A. Desmopression ✓

B. VWF concentrate

C. Tranexemic acid

D. Cryoprecipitate

E. FFPs

VIII XIII VWF

Ca D phenytoin

Carb SIADH

[illegible]

VIII XIII VWF

Ans: Desmopression

Explanation: Desmopression is associated with acute transient thrombocytopenia which is already a problem in type 2B, so Desmopression should be avoided in this subset of disease. All other options can be used for treatment.

91. Which of the following conditions is not associated with acquired von Willebrand syndrome?

A. [illegible]

AA

AL

AA

B. Chronic renal failure

C. Myeloproliferative disorder

D. MGUS

E. SLE

Ans: Chronic renal failure

Explanation: Acquired von Willebrand syndrome is a mild to moderate bleeding disorder that results from an acquired deficiency or dysfunction of von Willebrand factor. It has diverse pathology and may result from;

- Autoantibodies that impair von Willebrand factor function or increased its clearance
- Adsorption of high molecular weight von Willebrand factor multimers to malignant cells or platelets
- Proteolytic cleavage of von Willebrand factor after shear stress induced unfolding or decreased VWF synthesis.

Diseases that have been implicated include lymphoproliferative disorders and plasma cell disorders including monoclonal gammopathy of unknown significance, multiple myeloma and Waldenstorm macroglobulinemia, and autoimmune disorders; including SLE, scleroderma and antiphospholipid antibody syndrome. Aortic stenosis and ventricular septal defects which can trigger shear- induced conformational changes that increased von Willebrand factor proteolysis: thrombocytosis, including also leads to decrease VWF. Chronic renal insufficiency is typically associated with increased level of VWF.

92. A 23 year old male presented with uncontrolled bleeding after tooth extraction. Labs showed Hb 12.1, WBC 7000, and platelets 235,000. BT 12 minutes, PT 14 seconds and APTT 45 seconds. What is the probable diagnosis?

A. Hemophilia A

B. Hemophilia B

C. Thrombocytopenia

D. Glanzman thrombasthenia

E. Von Willebrand disease ✓

AS

Ans: von Willebrand disease

Explanation: Bleeding with normal platelets with increased BT and APTT is due to VWD.

93. Which of the following is the most common genetic cause of hemophilia A?

A. Frame shift mutation

B. Gene inversion ✓

C. Large deletion

D. Missense mutation

E. Nonsense mutation

50IUKg
25IU/kg 12.

Ans: Gene inversion

Explanation: Defects affecting the factor VIII gene is occurred due to many causes but the single most common and clinically important defect is an inversion involving F8 intron 22 which results 50% of all cases of severe disease.

94. A 13 year old boy, a diagnosed case of mild hemophilia A, presented with history of left hip pain for 4 days after fall from cycle. Examination showed pain on movement of left leg with hip flexed and thigh externally rotated. He also has paresthesia down the medial aspect of the thigh. Weight of patient is 35kg. How will you treat him?

A. 875 IU of recombinant FVIII

B. 1750 IU bolus followed by 875 IU every 12 hrs.

C. 1750 IU stat and then observe

D. Desmopression

E. No treatment needed

50IU/Kg
50
50
35
250 x
1750

Ans: 1750 IU bolus followed by 875 IU 12 hourly

Explanation: This hemophiliac patient has classical history of iliopsoas hematoma. This patient should be treated with FVIII concentrate at dose of 50 IU/kg bolus dose to correct the level to 100% followed by 25IU/kg 12 hourly until patient becomes asymptomatic, followed by 25IU/Kg every other day for a total of 14 days.

50IU/Kg

95. A 3 year old boy presented with uncontrolled bleeding after circumcision. Labs showed normal platelets with normal BT. PT is 14 s with APTT of 65. What is your diagnosis?

A. Von Willebrand disease

B. Vitamin K deficiency

[✓ C. Hemophilia]

D. ITP

E. Glanzman thrombasthenia

Ans: Excessive bleeding with high APTT is due to hemophilia.

96. A 72 year old female known case of RA presented with excessive vaginal bleeding for 1 month. There is no abnormality in genital tract. Labs showed normal platelets, BT, PT and fibrinogen with a prolonged APTT. Mixing studies do not correct the APTT. What is the likely diagnosis?

A. Congenital hemophilia A

B. Congenital hemophilia B

C. [✓ Acquired hemophilia]

D. Antiphospholipid antibody syndrome

E. Disseminated intravascular coagulation

aptt

APtt

~~XII~~

VIII

~~XII~~

Ans: Acquired hemophilia A

Explanation: The presence of an isolated prolonged aPTT suggests either an intrinsic pathway problem (deficient factor VIII) or a factor inhibitor. A mixing study must be ordered to determine whether there is an inhibitor; the results will show a correction of PTT if there is a factor deficiency, whereas the PTT will remain raised if a factor inhibitor is present. Isolated APTT is raised in congenital and acquired hemophilia, lupus anticoagulant and heparin therapy. Congenital hemophilia is less likely in a 72 years old female. Also, in congenital hemophilia APTT will be corrected with mixing studies. High APTT which does not correct with mixing is either due to acquired hemophilia or lupus anticoagulant. Lupus anticoagulant presents with thrombosis rather than bleeding. So, here RA patient with high APTT and excessive bleeding, the cause is acquired hemophilia. In addition, acquired hemophilia is sometimes mistaken for disseminated intravascular coagulation because both bleeding conditions are characterized by a prolonged aPTT. The following clinical characteristics distinguish the two diseases: prolonged PT, low fibrinogen, thrombocytopenia and elevated levels of fibrin degradation products and D-dimer.

Acquired hemophilia is a rare but potentially life-threatening bleeding disorder caused by the development of autoantibodies (inhibitors) directed against plasma coagulation factors, most frequently factor VIII. Most patients with acquired hemophilia present with hemorrhages into the skin, muscles, or soft tissues and mucous membranes. Intra-articular bleeding episodes are uncommon. The activated partial thromboplastin time typically shows a prolongation that is not reversed on a correction study. Reduced FVIII levels and evidence of an FVIII inhibitor are critical to the diagnosis of acquired hemophilia A. The following conditions may be associated with acquired hemophilia A :

- Pregnancy
- Autoimmune disorders (RA being most common)
- Inflammatory bowel disease
- Dermatologic disorders (e.g., psoriasis, pemphigus)
- Respiratory diseases (e.g., asthma, chronic obstructive pulmonary disease)
- Allergic drug reactions
- Diabetes
- Acute hepatitis B infection
- Acute hepatitis C infection
- Malignancies-solid tumors (prostate, lung, colon, pancreas, stomach, bile duct, head and neck, cervix, breast, melanoma, kidney)
- Hematologic malignancies

97. A diagnosed case of VWD patient needs tooth extraction. His dental surgeon needs your consultation. What will you advise?

VIII XIII VWF

A. FFP
B. Cryoprecipitate
C. VWD concentrate
D. Desmopression
E. No need of any treatment

2b Transm.

D
A W

Ans: Desmopression

Explanation: For minor surgical procedures and dental procedures, Desmopression can be used to increase the VWF level before surgery, and it is recommended to use it in all patient with or without Tranexemic acid before giving VWF concentrate.

98. A 10 year old male presented with recurrent epistaxis. Investigations show normal platelets with BT of 11 minutes (increased). PT and APTT are normal. Further testing shows decrease primary platelets aggregation response with normal response to ristocetin. What is your diagnosis?

A. VWD variant
B. Storage pool disease
C. Glanzman thrombasthenia

D. Hemophilia

E. ITP

Ans: Glanzman thrombasthenia

Explanation: Glanzmann thrombasthenia (GT) is a rare genetic platelet disorder in which the platelets have qualitative or quantitative deficiencies of the fibrinogen receptor αIIbβ3. When an injury occurs, the αIIbβ3 receptors play an important role in the adherence of platelets to the endothelium as well as have a role in platelet aggregation.

The αIIbβ3 complex binds fibrinogen and/or von Willebrand factor. Adjacent platelets are cross-linked through the αIIβ3. When the complex functions abnormally, platelets cannot aggregate. This then leads to increased bleeding. It presents with mucosal bleeding. Labs showed normal platelets with increased bleeding time. PT and APTT are normal. On platelet aggregation studies, the primary platelet aggregation response to platelet agonists such as adenosine diphosphate (ADP), epinephrine, and collagen are decreased, whereas the response to ristocetin is normal; *if the secondary platelet aggregation response is abnormal, suspect a platelet storage pool defect or an abnormality in platelet signal transduction.*

99. A 28 year old female presented with sudden onset of shortness of breath and pleuritic chest pain for 3 hours. Patient is 3 week post-partum after C section at 36 weeks due to pre eclampsia. Chest X-RAY is normal. What is the preferred imaging modality in this patient to confirm the suspected diagnosis?

    A. CTPA
    B. MRI chest
    C. V/Q scan ✓
    D. Echocardiography
    E. Conventional angiography

V/Q

2/3

3-4

Aspirin | DVT Antipho

Ans: V/Q scan

Explanation: V/Q scan is preferred over CTPA when the baseline Chest X-RAY is normal, as it limits radiation exposure to the mother breast tissue.

100. A 24 year old woman with a history of DVT 3 years back while on OCPs in now 9 weeks pregnant. Her obstetrician wants your recommendation regarding antithrombotic recommendation. What will you recommend?

A. Monitor

B. Prophylactic LMWH

C. Rivaroxaban

D. Therapeutic LMWH

E. Warfarin with INR 2-3

Ans: Prophylactic LMWH

Explanation: Guidelines recommend prophylactic LMWH during pregnancy in a patient with history of DVT related to estrogen therapy. Warfarin is contraindicated in 1st trimester. DOAC are not recommended in pregnancy.

101. A patient with a history of recurrent thromboembolic events develops a DVT despite full anticoagulation with heparin. Which one of the following causes of thrombophilia is associated with resistance to heparin?

Ans

A. Protein C deficiency

B. Protein S deficiency

[✓ C. Antithrombin III deficiency]

D. Lupus anticoagulant

E. Homocystienuria

Ans: Antithrombin III deficiency

Explanation: Antithrombin III (ATIII) is a nonvitamin K-dependent protease that inhibits coagulation by neutralizing the enzymatic activity of thrombin (factors IIa, IXa, Xa). Antithrombin III activity is markedly potentiated by heparin, the principal mechanism by which both heparin and low–molecular-weight heparin result in anticoagulation. Antithrombin III deficiency is an autosomal dominant disorder which predispose to thrombosis. Because antithrombin is not present, anticoagulation with heparin is unreliable in this thrombophilia. Once a patient with congenital antithrombin III deficiency has developed thrombosis, anticoagulation is more strongly indicated. Warfarin is the principal anticoagulant used. This vitamin K antagonist is administered at a dose to maintain an international normal ratio of 1.5-2.5.

102. A newborn baby has continuous bleeding from umbilical cord stumps. His two other siblings have the same problem. Which test you will order for him?

A. PT

B. [✓ APTT]

C. Bleeding time

D. CBC

E. D dimers

Ans: APTT, family history of bleeding think about Hemophilia A, do APTT.

103. A 40 year old female diagnosed case of stage 3 breast carcinoma on chemotherapy presented with acute painful right leg swelling. Doppler USG confirms the diagnosis of deep vein thrombosis. What should be your management plan for her?

A. Give LMWH for 5 days then switch to warfarin for 6 months

B. Give LMWH for 5 days then switch to rivaroxaban for 6 months

C. Observe only

D. [✓ Subcutaneous LMWH for 6 months]

E. Compression stocking only

DVT

P.

Ans: Subcutaneous LMWH for 6 months

Explanation: Venous thromboembolism (VTE) is a complex and common complication of cancer and its treatment. Compared with VTE in patients without cancer, cancer-associated VTE is associated with higher rates of recurrence, bleeding complications associated with anticoagulation therapy and mortality. LMWH is the preferred agent for the initial and long-term treatment of VTE in patients with cancer. The treatment duration is mostly 6 months but patient may need lifelong anticoagulation. Treatment with warfarin in cancer patients is associated with higher risk of bleeding and more risk of recurrence. DOAC are not recommended but some studies show benefit if cost is an issue.

104. A female presented with sudden onset of chest pain with SOB. She was taking warfarin for an above knee DVT for last 15 days .On examination patient has HR of 150/ min with BP of 80/40 mmhg. ECG showed sinus tachycardia and S1Q3T3 pattern with right ventricular strain. What is the next best management?

A. Enoxaparin 1mg/kg twice daily

B. [✓ Unfractionated heparin]

C. Continue warfarin

D. Thrombolysis

E. Shift to Rivaroxaban

Ans: Thrombolysis

Explanation: This is a case of massive pulmonary embolism, the treatment of which is urgent thrombolysis. Massive PE is defined as PE causing hemodynamic compromise.

105. A 25 year old lady presented with a swollen left lower limb when she was 12 weeks pregnant. Doppler studies confirmed a femoral vein DVT. There was no family history of thrombophilia. What is the management of this patient?

A. LMWH throughout pregnancy, warfarin 6 weeks postpartum

B. Consider termination of pregnancy

C. Compression stocking during pregnancy, warfarin 6 weeks postpartum

D. Warfarin throughout pregnancy and 6 weeks postpartum

E. Aspirin during pregnancy and warfarin 6 weeks postpartum

Ans: LMWH throughout pregnancy and warfarin 6 weeks postpartum

Explanation: Pregnancy increases the risk of venous thromboembolism (VTE) 4- to 5-fold over that in the nonpregnant state. Subcutaneous low molecular weight heparin (LMWH) is the preferred treatment for most patients with acute VTE and it should be given throughout the pregnancy. Warfarin should never be used for DVT treatment in pregnancy because it is teratogenic, the only exception is in the presence of prosthetic heart valves needing anticoagulation for which low dose warfarin can be used during 2nd and 3rd trimester. Pregnant patients with acute VTE are typically treated with therapeutic anticoagulation for a minimum of 6 months, and for at least 6 weeks postpartum. VTE is more common in the postpartum period. Postpartum therapeutic anticoagulation may consist of continued use of therapeutic heparin, or the patient may be bridged to warfarin. Discontinuation of heparin early in the transition to warfarin may cause increased short-term risk of VTE. To minimize this risk during conversion to warfarin, the patient should remain on therapeutic anticoagulation with heparin for at least 4-5 days while the warfarin is titrated to a goal INR of 2.0-3.0.

106. An 8 year old boy who is diagnosed case of severe hemophilia A came with muscle hematoma. Factor VIII concentrate was not available. What blood component will you transfuse?

A. Whole blood

B. FFPs
C. Platelets
D. Cryoprecipitate
E. PRBCs

VIII XIII. VWF.
Fibrino

Ans: Cryoprecipitate

Explanation: For Hemophilia, if FVIII concentrate not available, Cryoprecipitate can be used as it contains high level of factor VIII.

107. Which one of the following thrombophilia predisposes to both arterial and venous thrombosis?

A. Factor V Laden mutation
B. Antithrombin III deficiency
C. Protein S deficiency
D. Antiphospholipid antibody syndrome ✓
E. Dysfibrinogenemia

5-

VIII
XIII VWF

Ans: Antiphospholipid antibody syndrome

Explanation: Antiphospholipid antibody syndrome and Homocystienuria predispose to both arterial and venous thrombosis. All other thrombophilia increased the risk of venous thrombosis.

108. A patient is started on warfarin for unprovoked DVT 4 days back. Patient has developed purpura of skin, INR is 1.9. What is the cause of this problem?

A. Protein S deficiency
B. Protein C deficiency ✓
C. Antithrombin III deficiency
D. Factor V Laden mutation
E. Prothrombin gene mutation

II

Ans: Protein C deficiency

Explanation: Skin reactions associated with warfarin commonly occur 3 to 5 days after initiating the treatment. Breasts, buttocks, abdomen, thighs, and calves are more susceptible probably because of the reduced blood supply to adipose tissues. Rapid progression of plaques to skin necrosis increases morbidity. Skin necrosis usually occurs shortly after initiating warfarin therapy with a large loading dose or without concomitant heparin. Warfarin inactivates vitamin K-dependent clotting factors II, VII, IX, and X. At the same time, vitamin K-dependent proteins C and S are inactivated. This may cause a paradoxical hypercoagulable milieu in which micro thrombi develop in cutaneous and subcutaneous venules, as the concentration of anticoagulant protein C falls more rapidly than other vitamin K-dependent procoagulant factors, which have longer half-lives. Protein C is a Vit K dependent anticoagulant with a half-life of 6 hours. It is converted to activated protein C by thrombin. Activated protein C in combination with protein S degrades Va and VIIIa. It also binds to PAI-1 and enhance fibrinolysis. Heterozygous deficiency is present in 0.3% of population and can leads to VTE. Skin reaction are common in its deficiency at the start of warfarin therapy.

109. Which of the following test is the gold standard for the diagnosis of DVT?

A. Duplex ultrasound
B. D- Dimer
C. CT angiography
D. Contrast venography
E. [✓ MR venography]

CTPA

Careful

Ans: Contrast venography

Explanation: First test=> Duplex ultrasound, it is called duplex because of two parts of the test. In the first part of the process a B mode ultrasound is used to obtain an image through sound waves that are bounced back and make echoes. During this process, compression done. If vein cannot be compressed, a DVT diagnosis is made. The ability to completely flatten a vein with compression rules out DVT. In the second part of process, blood flow abnormalities are detected. Absence of blood flow confirms DVT. Duplex Ultrasound has 95% sensitivity for above leg DVT and 75% sensitivity for below leg DVT. It cannot detect isolated pelvis vein thrombosis.

Gold standard test is contrast enhanced venography. MR venography can also be used but it's commonly used when patients have plaster casts. D – Dimer is used for its negative predictive value.

110. A 7 year old girl requiring a tonsillectomy undergoes pre-operative laboratory testing which revealed a prolonged APTT of 80 seconds. Prothrombin time is normal. Patient has no history of any bleeding. Mixing with plasma corrected the APTT. What is the diagnosis?

A. Hemophilia A
B. Hemophilia B

C. Factor VII deficiency

D. Antiphospholipid antibody syndrome

E. Factor XII deficiency

Ans: Factor XII deficiency ✓

Explanation: Prolonged APTT which gets corrected by plasma means deficiency of intrinsic pathway factors and not the inhibitors. Hemophilia A and B are X- linked disorders and not occur in female except in Turner syndrome. Also patient has no history of bleeding which also rules out Hemophilia. Factor VII deficiency would increase PT not APTT. Factor XII deficiency can cause an increase in APTT without bleeding. Factor XII deficiency is a rare genetic blood disorder that causes prolonged clotting (coagulation) of blood in a test tube without the presence of prolonged clinical bleeding tendencies. It is caused by a deficiency of the factor XII (Hageman factor). It is sometimes associated with thrombosis rather than bleeding. Factor XII deficiency has not been linked to any significant hemorrhagic diatheses. The disorder may be considered in patients with prolonged aPTT, normal PT, normal bleeding time, and no clinical history of bleeding. Once thought likely, the deficiency can be confirmed by normalization of aPTT with plasma component therapy and by factor assay.

111. Which of the following type of lymphoma is most likely to have CNS involvement?

A. Burkitt Lymphoma

[✓ B. Diffuse large B cell lymphoma]

C. Hodgkin's lymphoma

D. Lymphoplasmacytic lymphoma

E. Mantle cell lymphoma

3

2

Ans: Diffuse large B cell Lymphoma

Explanation: Rate of CNS involvement is more with aggressive type of non-Hodgkin's Lymphoma, like diffuse large B cell lymphoma and T cell lymphoma.

112. A 25 year old female presented with left calf pain and swelling for 4 days. Duplex ultrasound of leg confirmed popliteal vein thrombosis. Patient was admitted in hospital for cholecystectomy 10 days before. What should be next treatment for her?

A. Anticoagulation for 6 months

B. Anticoagulation for 3 months ✓

C. Lifelong anticoagulation

D. LMWH for 1 week followed by compression stocking

E. Aspirin and compression stocking

Ans: Anticoagulation for 3 months

Explanation: This is a case of provoked DVT. Guidelines recommend 3 months anticoagulation for provoked DVT and 6 months for unprovoked DVT. Aspirin can be used for secondary prevention of DVT.

113. How much units of unfractionated heparin for 24 hours is required to achieve full anticoagulation?

A. 25000 IU

B. 30,000 IU

C. 35000 IU ✓

D. 40000 IU

E. 50,000 IU

35

25,000

5000

4cc

Ans: 35000 IU of unfractionated heparin is required in 24 hours' time to achieve full anticoagulation.

114. Which one of the following is the side effect of long term administration of heparin?

A. Thrombocytopenia

B. Bleeding

C. Increased ALT.

D. Osteoporosis ✓

E. Anaphylaxis

Ans: Osteoporosis

Explanation: Osteopenia and Osteoporosis is a side effect of long term heparin use, which is mostly used in pregnancy and for patients with cancer and DVT. All other side effects occur within 2 weeks of therapy except osteoporosis.

10cm

115. A 67 year old female is referred to you for consultation after an abnormal CBC result detected on routine annual screening which showed Hb 12.3, WBC of 29,000 with 85% lymphocytes and platelets 235,000. Peripheral film done which showed small lymphocytes with smudge cells. Patient is otherwise asymptomatic with no history of fever or weight loss. Examination showed cervical lymphadenopathy and mild splenomegaly. What is the next best treatment for her?

A. Chlorambucil therapy

B. Fludarabine therapy

C. Rituximab therapy

D. Ibrutinib therapy

E. Observe with regular follow up

FCR

[illegible] R

Ans: Observe with regular follow up

Explanation: This patient is having CLL as manifested by high WBC counts with predominant small lymphocytes and smudge cells with lymphadenopathy and splenomegaly. As patient is asymptomatic, treatment is not indicated. Indication of treatment in CLL are:

- Development of anemia/+ thrombocytopenia due to marrow failure
- Massive (>10cm) or progressive lymphadenopathy
- Massive or progressive splenomegaly
- Progressive lymphocytosis with >50% increase in 2 months or lymphocytes doubling time < 6 months
- Systemic symptoms with weight loss >10% in previous 6 months, fever >38 for more than 2 weeks, extreme fatigue and drenching night sweats
- Autoimmune hemolytic anemia or thrombocytopenia

116. An 18 year old man presented with recurrent epistaxis, hemoptysis and two episodes of melena. Blood tests including coagulation parameters are within normal limits except low Hb. CXR showed multiple pulmonary nodules. Patient has multiple dilated veins on lips and nose. What is the most likely diagnosis?

A. Acute lymphocytic leukemia

B. Cancer associated bleeding

C. Osler Weber rendu syndrome

D. Peutz Jaghers syndrome

E. Ehler Danlos syndrome

10

HHT

Ans: Osler Weber Rendu syndrome

Explanation: Diagnostic criteria for Osler Weber Rendu syndrome also called hereditary hemorrhagic telangiectasia is;

- Recurrent epistaxis
- Telangiectasia, at multiple sites like lips, oral cavity, fingers and nose.
- Visceral lesions like GI telangiectasia causing recurrent bleeding, pulmonary AV malformation, hepatic AV malformation, cerebral AVM, spinal AVM
- First degree relative with the disease

If 2 of the 4 present, probable diagnosis. If 3 or more than 3, definite HHT.

117. A 43 year old woman presented with heaviness and aching in the right leg. Patient is on warfarin therapy for proximal DVT for 4 months. INR is 2.7. Patient complains of increased swelling and itching in the calf region. How will you treat this patient?

A. Continue treatment and review after 2 months
B. Increase the dose to target INR 3.5
C. Shift to heparin
D. Compression stocking
E. Thrombolysis

2.5

3.5

3.0

Ans: Compression stocking

Explanation: The presence of swelling with itching is suggestive of post thrombotic syndrome. Venous outflow obstruction and venous insufficiency result in chronic venous hypertension resulting in post thrombotic syndrome. Features are painful heavy calves, pruritus, swelling and varicose veins. Compression stockings should be offered to all patients with DVT to prevent this complication.

It is less likely that patient on therapeutic INR develops new DVT.

118. In which of the following anemia, splenectomy is most helpful?

A. Hereditary spherocytosis
B. Thalassemia major
C. Sickle cell disease
D. G6PD deficiency
E. Autoimmune hemolytic anemia

10,000

30,000

Py

I

IVIG

Ans: Hereditary spherocytosis => splenectomy almost corrects the anemia of HS.

119. Which of the following organism has the highest risk of sepsis after splenectomy?

A. Pneumococcus

B. Heamophilus influenza

C. Meningococcus

D. Malaria

E. Bebesiosis

Ans: Pneumococcus

Explanation: Although the risk of infection of all the above organisms is Increased after splenectomy, but pneumococcus imposes the highest risk of sepsis with 7% risk per year. Sepsis due to pneumococcus is also severe with high mortality in patients with no spleen. That is why it is the most important vaccine in splenectomized patients and should be done 2 weeks prior to surgery and after 5 years.

120. A patient has developed DVT. Labs were done which showed PT 15 second and APTT of 69 seconds. Mixing studies were done which does not correct the APTT. What is the most probable diagnosis?

A. Protein C deficiency

B. Phospholipid antibody syndrome

C. Factor VIII antibodies

D. Antithrombin III deficiency

E. DIC

XII VIII XIII VIII XII XIII

Oups

Ans: Anti phospholipid antibody syndrome

Explanation: If patient has raised APTT, the next test is mixing studies in which patient plasma is mixed with normal plasma containing all the factors. If APTT gets corrected with mixing it showed factor deficiency for which individual factor levels should be done. If APTT does not correct with mixing, there are three possibilities. *Lupus anticoagulant, presence of inhibitors of coagulation factors and DIC.* Lupus anticoagulant presents with recurrent arterial and venous thrombosis, factor inhibitors present with bleeding. In DIC, PT will also be raised.

121. Which of the following thrombophilia increased the risk of developing VTE in patients taking oral contraceptive pills?

A. Antithrombin III deficiency ✓

B. Protein C deficiency

C. Protein S deficiency

D. Factor V Laden mutation ✓

E. Thrombin gene mutation

Ans: Factor V laden mutation

Explanation: Healthy women taking OCPs have a 3- to 4-fold increased risk of developing a DVT or PE compared with women who do not take OCP. Women with factor V Leiden who take OCPs have about a 35-fold increased risk of developing a DVT or PE compared with women without factor V Leiden and those who do not take OCPs. This would translate to an about 35 per 10 000 chance per year of use on average for women in their twenties with factor V Leiden. Likewise, postmenopausal women taking HRT have a 2- to 3-fold higher risk of developing a DVT or PE than women who do not take HRT, and women with factor V Leiden who take HRT have a 15-fold higher risk. This is about a 15 to 40 per 1000 chance per year of use, on average, for women in their fifties with factor V Leiden. Women with heterozygous factor V Leiden who are making decisions about OCP or HRT use should take these statistics into consideration when weighing the risks and benefits of treatment.

122. A patient with trauma with massive bleeding came to surgical emergency. The surgical resident needs your help regarding transfusion in emergency without cross matching. Which of the following is the best to give?

[✓ A. O-ive]
B. O+ ive.
C. AB +ive
D. AB – ive
E. A + ive

ALL.

6cm:
60kd

10
0cm:
6cm
barrow
β-blasts

Ans: O – ive

Explanation: Individuals with blood group O – ive are called universal donor and in emergency condition where full cross match cannot be done, this can be transfused safely. Blood group AB can only be given to AB group only. O + ive blood will sensitize the O – ive recipient.

123. A 32-year-old woman from South Punjab was admitted to hospital with shortness of breath and fatigue. Physical examination showed massive splenomegaly and pallor. Further investigations showed low hemoglobin and raised IgM level. What is the probably diagnosis?

A. Leishmaniasis
[✓ B. Tropical splenomegaly]
C. CML

D. Myelofibrosis

E. Walderstorm macroglobulinemia

Ans: Tropical splenomegaly

Explanation: Tropical splenomegaly syndrome, is a poorly understood condition thought to represent an exaggerated immune response to recurrent or persistent malarial infection. Tropical splenomegaly syndrome is characterized by a polyclonal hypergammaglobulinemia and raised immunoglobulin M (IgM). Criteria for the diagnosis of tropical splenomegaly syndrome are based on 1) the exclusion of other causes of splenomegaly; 2) immunity to malaria, defined as a strongly positive antibody test; 3) splenomegaly of at least 10 cm; 4) a serum concentration of IgM at least two standard deviations above normal; and 5) a clinical and immunological response to malaria prophylaxis.

6cm

IgM

IgM

ATT

IRS

4.6